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Juvenile Fibrosarcoma of the Temporal Bone

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A case of juvenile fibrosarcoma arising from the head and neck region is described. This type of tumour should be considered as a separate entity different from the fibrosarcoma in adults because of the different clinical behaviour. The symptomatology, the radiographic features and the literature data are reviewed. © 1996 Wiley-Liss, Inc.

Key words: fibrosarcoma, magnetic resonance imaging, temporal bone

CASE REPORT

A two-year-old girl was referred to our hospital for evaluation of a left sided preauricular mass. On examination the skin overlying the mass was normal although an involuting tuberous hemangioma was seen anteriorly. The mass itself was hard, painless, and immobile. Further clinical examination was normal. Full blood count and biochemistry showed no abnormality. Tumour markers were negative.

On brain computed tomography (CT) a space occupying lesion was visible in the left temporal bone. There was invasion of the diploic space with ballooning and destruction of both the inner and outer table (Fig. 1). The mass extended to the petrous bone. Brain magnetic resonance imaging (MRI) confirmed the extracerebral localisation of the tumour but demonstrated its close relationship to the dura (Fig. 2). On CT and MRI there was peripheral enhancement of the lesion and digital subtraction angiography (DSA) confirmed a moderate predominantly peripheral vascularisation after selective injection of the external carotid artery. Two tuberous hemangiomas were detected.

A biopsy was taken and microscopical examination revealed tumoral spindle cells with a variable amount of cytoplasm and an increased mitotic activity (Fig. 3). Large areas of necrosis were noted. Immunohistochemical examination showed positivity for vimentin and α smooth muscle actin and negativity for prekeratin and desmin. The diagnosis of a mixed sarcoma with characteristics of both fibrosarcoma and leiomyosarcoma was put forward.

Although total excision of the tumour would have been possible, an "en-bloc" resection with wide margins was not an acceptable option in this localisation. The child was treated with chemotherapy (ifosfamide, vincristine, dactinomycin) according to the SIOP (Société Internationale d'Oncologie Pédiatrique) protocol for malignant mesenchymal tumours (MMT '89)

Clinically there was no further increase in the size of the tumour. After six courses of chemotherapy the tumour was totally excised. Microscopical and immunohistochemical examination showed similar findings except for the absence of necrosis and mitotic activity. The tumour no longer had malignant cytological characteristics and this was considered to be due to the effect of chemotherapy. At the ultrastructural level, the tumour cells showed features of myofibroblasts.

There was no evidence of residual tumour on a brain CT 11 months after surgery. Eighteen months after the treatment, the child is clinically extremely well.

DISCUSSION

Malignancies involving the head and neck region are rare and account only for 5% of the childhood cancers [1]. The major malignancies of this region are lymphoma (59%), rhabdomyosarcoma (13%), and thyroid carcinoma (10%). Nasopharyngeal carcinoma and neuroblastoma (5%), salivary gland carcinoma (2.5%), and malignant teratoma (1%) are less common. In the Pittsburgh study (241 cases), fibrosarcoma accounted only for 0.4% [2].

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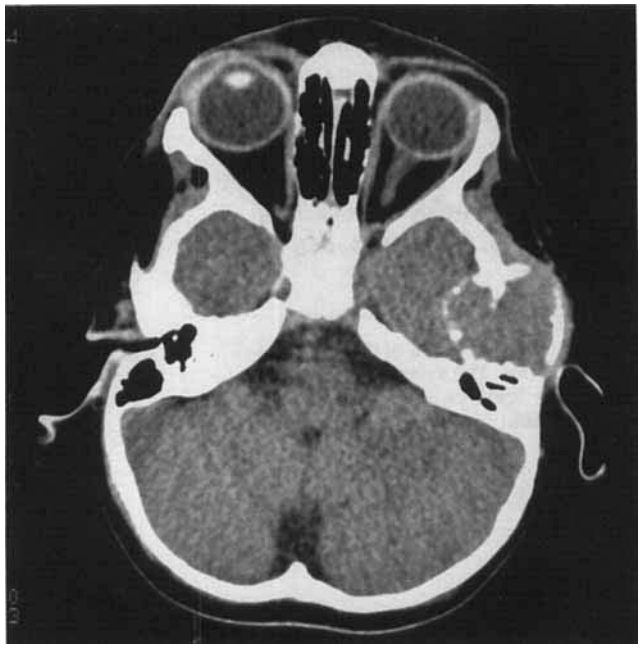

Fig. 1. Unenhanced brain CT. A mass is seen in the temporal bone with expansion and erosion of the inner and outer table.

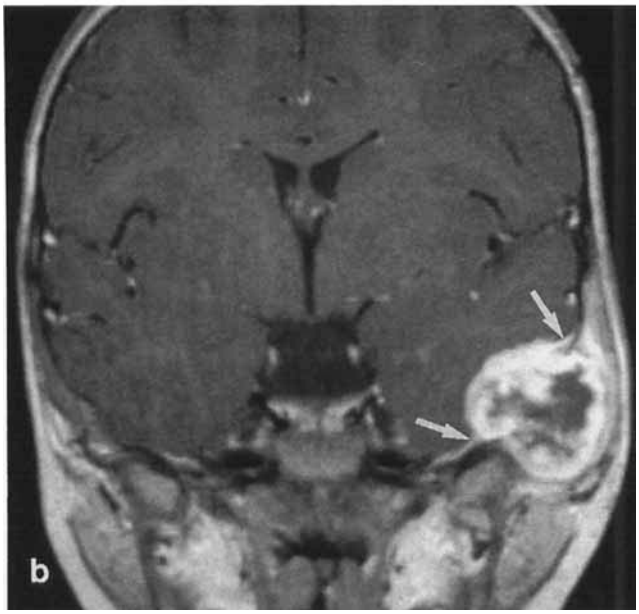

Fig. 2. Axial T_2 weighted (TR/TE = 2500/90) **(a)** and Coronal T1 weighted SE sequence (TR/TE = 520/15 ms) **(b)** after gadopentetate dimeglumine enhancement. The tumour returns a low signal on T1 weighting (b). There is a strong peripheral enhancement (b). The close relationship with the dura is shown (b, arrows).

More than 50% of the infantile fibrosarcomas are diagnosed in the first year of life. Approximately one third are present at birth (congenital fibrosarcoma) [3]. More than 70% of the tumours originate in the extremities and less than 2% in the head and neck region [4–7]. There is a slight male preponderance [5]. The 5-year survival rate of the infantile fibrosarcoma is 84 to 92% [5]. Local lymph node involvement and metastases in children occur in less than 10% of the cases compared to 50% in adults [8].

Childhood fibrosarcoma is histologically somewhat different from the adult tumour. The cells look more primitive, more rounded, and less pleomorphic.

Radiologically fibrosarcoma appears as a non-specific soft tissue tumour. Adjacent bone infiltration with destruction and calcification may occur. Particularly in children, severe erosion and bone destruction can be seen. Therefore CT is the technique of choice in the diagnostic work-up. MRI can be of help in assessing dural invasion. On arteriography, fibrosarcoma is a hypervascular tumour usually with marked neovascularisation. Because of the non-specific characteristics, there is a large differential diagnosis including rhabdomyosarcoma, leiomyosarcoma, malignant fibrous histiocytoma, malignant nerve sheath tumour, chondrosarcoma, malignant mesenchymoma (mixed rhabdomyosarcoma and chondrosarcoma) [9].

Treatment for fibrosarcoma in both adults and children is usually surgery. Since local recurrence may occur in up to 40% of cases after surgical resection, wide "en-bloc" resection is preferred [8,10]. If this is impossible due to anatomical localisation or because it requires mutilating surgery, preoperative chemotherapy might be appropriate in infants and children [11,12].

The rationale is that after chemotherapy more conservative surgery is justified. This can be extremely important in young children and infants where "en-bloc" resec-

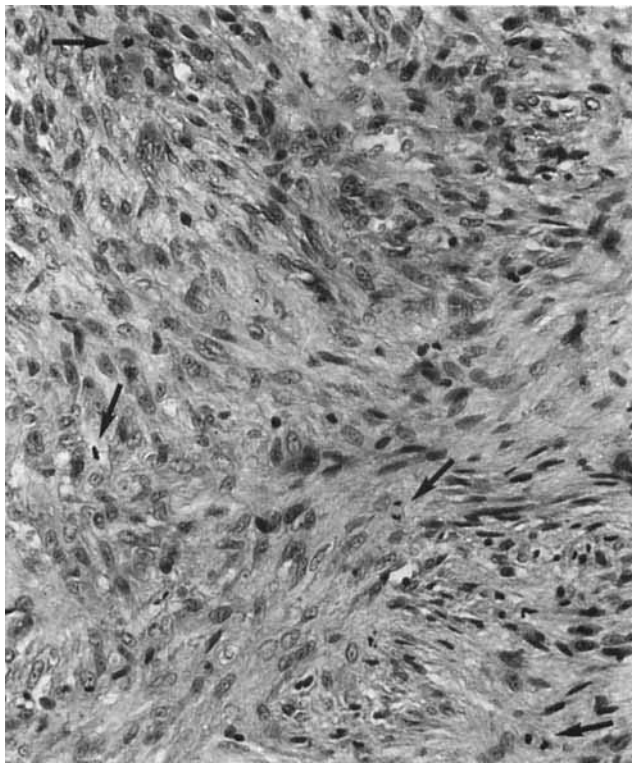

Fig. 3. The tumour was composed of plump and somewhat bundled spindle cells showing increased mitotic activity (arrows) (hematoxylin and eosin stain, ×312).

tion may carry a high morbidity. Children with fibrosarcoma under the age of 1 year (infants) are excluded from the MMT '89 protocol, as they have an extremely good prognosis. Even spontaneous tumour regression has been reported in this age group [13]. Infants should probably receive a less aggressive chemotherapy regimen followed by conservative surgery [14].

REFERENCES

1. Snyderman NL, Smith RJH: Controversies: fibrosarcoma of the infratemporal fossa in a 8 year old girl. Head Neck 13:156–159, 1991.
2. Cunningham MJ, Myers EN, Bluestone C: Malignant tumors of the head and neck in children; a 20 year review. Int J Pediatr Otorhinolaryngol 13:279–292, 1987.
3. Pinto A, Dold OR, Mueller D, Gilbert-Barness E: Pathological cases of the month: infantile fibrosarcoma. Am J Dis Child 147: 691–692, 1993.
4. Bang G, Baardsen R, Gilhuus-Moe O: Infantile fibrosarcoma in the mandible: case report. J Oral Pathol Med 18:339–343, 1989.
5. Chung EB, Enzinger FM: Infantile fibrosarcoma. Cancer 38:729–739, 1976.
6. Hayani A, Mahoney DH, Hawkins HK, Steuber CP, Hurwitz R, Fernbach DJ: Soft-tissue sarcomas other than rhabdomyosarcoma in children. Med Pediatr Oncol 20:114–118, 1992.
7. Singh PK, Singh RK, Agarwal A, Rajvanshi VS: Fibrosarcoma of the middle ear. Ear Nose Throat J 68:479–480, 1989.
8. Ninane J, Gosseye S, Paneon E, Clause D, Rombouts J-J, Cornu G: Congenital fibrosarcoma. Cancer 58:1400–1406, 1986.
9. Paulus W, Slowik F, Jellinger K: Primary intracranial sarcomas: histopathological features of 19 cases. Histopathology 18:395–402, 1991.
10. Mark RJ, Sercarz JA, Tran L, Selch M, Calcaterra TC: Fibrosarcoma of the head and neck. The UCLA experiment. Arch Otolaryngol Head Neck Surg 117:396–401, 1991.
11. Brock P, Renard M, Smet M, De Wever I, Casteels-Van Daele M: infantile fibrosarcoma. Med Pediatr Oncol 19:210, 1991.
12. Ninane J, Rombouts JJ, Cornu G: Chemotherapy for infantile fibrosarcoma. Med Pediatr Oncol 19:209, 1991.
13. Madden NP, Spicer RD, Allibone EB, Lewis IJ: Spontaneous regression of a neonatal fibrosarcoma. Br J Cancer 66 (suppl XVIII): S72–S75, 1992.
14. Spicer RD: Chemotherapy for infantile fibrosarcoma. Med Pediatr Oncol 21:80, 1993.